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SHORT COMMUNICATION

Case report

Bilateral central anterior stromal opacity of the cornea: dystrophy or degeneration?

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PURPOSE. *To describe a bilateral, central, oval corneal opacity not resembling any known corneal degeneration or dystrophy.*

METHODS. *Ophthalmic examination, corneal topography, and pachymetry.*

RESULTS. *A 30-year-old woman presented complaining of blurred vision. Biomicroscopic examination revealed bilateral, centrally located, oval, diffuse opacification of the anterior corneal stroma. Corneal topography showed slightly paracentral corneal flattening and irregular astigmatism in both eyes. Pachymetry indicated thinning of the central cornea in the left eye. No evidence of systemic disease was found. Family members had no corneal abnormalities.*

CONCLUSIONS. *This unusual keratopathy closely resembles climatic proteoglycan stromal keratopathy. However, it is not clear whether the condition should be described as a dystrophy or degeneration. (Eur J Ophthalmol 2003; 13: 315-9)*

KEY WORDS. *Anterior stromal opacity, Climatic proteoglycan dystrophy, Corneal thinning, Corneal flattening*

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INTRODUCTION

A number of dystrophies and degenerations involving the central cornea have been described that may have various impacts on visual acuity (1). Rare and subtle conditions of the cornea are infrequently reported but can provide valuable insights into diseases of the cornea.

Here we describe bilateral oval opacification of the central corneal stroma and central corneal flattening and thinning as isolated findings in a young, healthy patient with no history of ocular or systemic disease.

Case report

A 30-year-old woman presented, complaining of blurred vision. She had been wearing eyeglasses for ten years and mentioned that her prescription had changed several times within the last two years. Visual acuity was 9/10 in the right eye (RE) and 8/10 in the left (LE), with a manifest refraction of RE +0.25 +1.50 x 10 and LE +1.25 +5.00 x 175. Biomicroscopic examination showed bilateral, centrally located, oval, gray diffuse opacification of the corneal stroma and epithelial iron deposits over the lesion (Fig. 1). Narrow slit illumination indicated that this haze mainly occupied the anterior stroma and was denser at the level of Bowman membrane (Fig. 2). The tear film and endothelial layer were unremarkable; and the ocular surface did not stain with fluorescein. The remainder of the anterior segment including the iridocorneal angle was normal. Corneal sensitivity measured with the Cochet-Bonnet esthesiometer was normal. Corneal topography showed slightly paracentral corneal flattening and irregular astigmatism in both eyes (Fig. 3). Orbscan pachymetry indicated central corneal thickness was 525 µm in RE and 451 µm in LE (Fig. 4).

The patient had no history of systemic disease, ocular trauma, or infection. She was not taking any sys-

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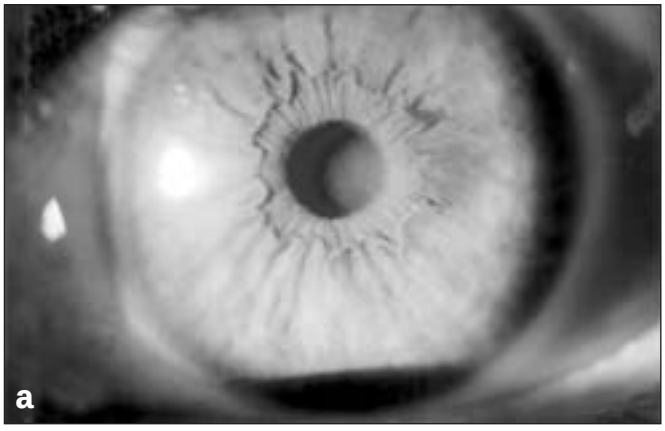

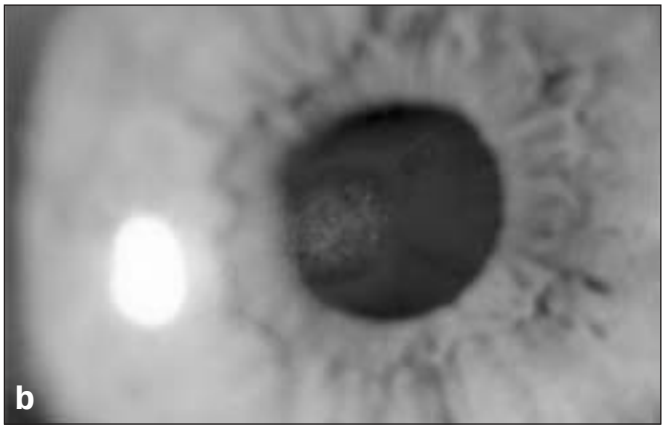

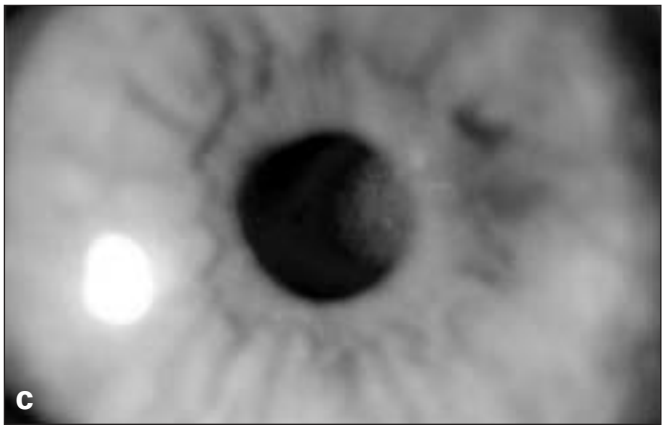

Fig. 1 - *An oval area of gray stromal haze and epithelial iron deposits in the central cornea of left (**a**, **c**) and right (**b**) eyes.*

temic medications. Hematologic examination and lipoprotein profile (very low-density, low-density, and high-density lipoprotein, triglycerides, cholesterol, apolipoprotein I and II) were normal. Serologic tests for syphilis (Venereal Disease Research Laboratory and

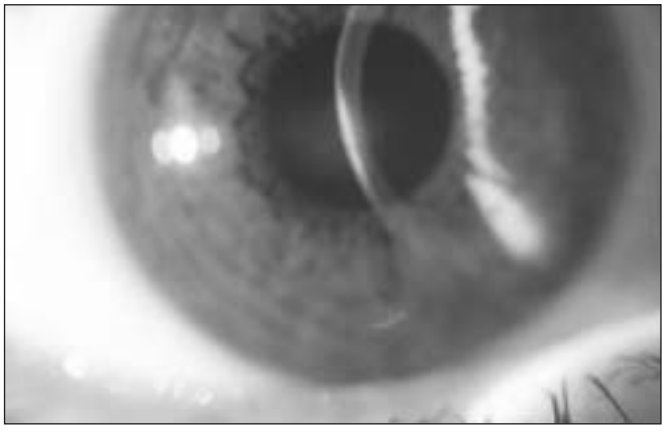

Fig. 2 - *Narrow slit illumination indicates that the opacity occupies the anterior corneal stroma.*

Treponema pallidum hemagglutination) were negative. Family members (parents and four sisters) were examined and none of them had any similar corneal disorder.

DISCUSSION

Corneal dystrophies with central corneal opacity associated with corneal flattening and thinning that resemble the keratopathy in our patient are few. One of them is macular corneal dystrophy without the macules, which starts with a fine superficial stromal haze involving the central cornea during the first decade of life. Central corneal thickness may be significantly reduced in macular dystrophy (2). However, the opacification extends to the periphery and involves the entire thickness of the stroma by the second decade, leading to marked loss of vision. Macular dystrophy is inherited as an autosomal recessive trait but it may seem sporadic because the heterozygous carriers do not present corneal changes.

Posterior amorphous corneal dystrophy is a rare autosomal dominant disease characterized by stromal opacification and corneal flattening and thinning (3, 4). Gray sheet-like opacities are most prominent in the deeper stromal layers and the opacification begins at a young age. A similar corneal opacity was first reported by Waring et al (5). They described bilateral corneal degeneration in 29 patients, with central, horizontally oval or round, gray ground-glass haze

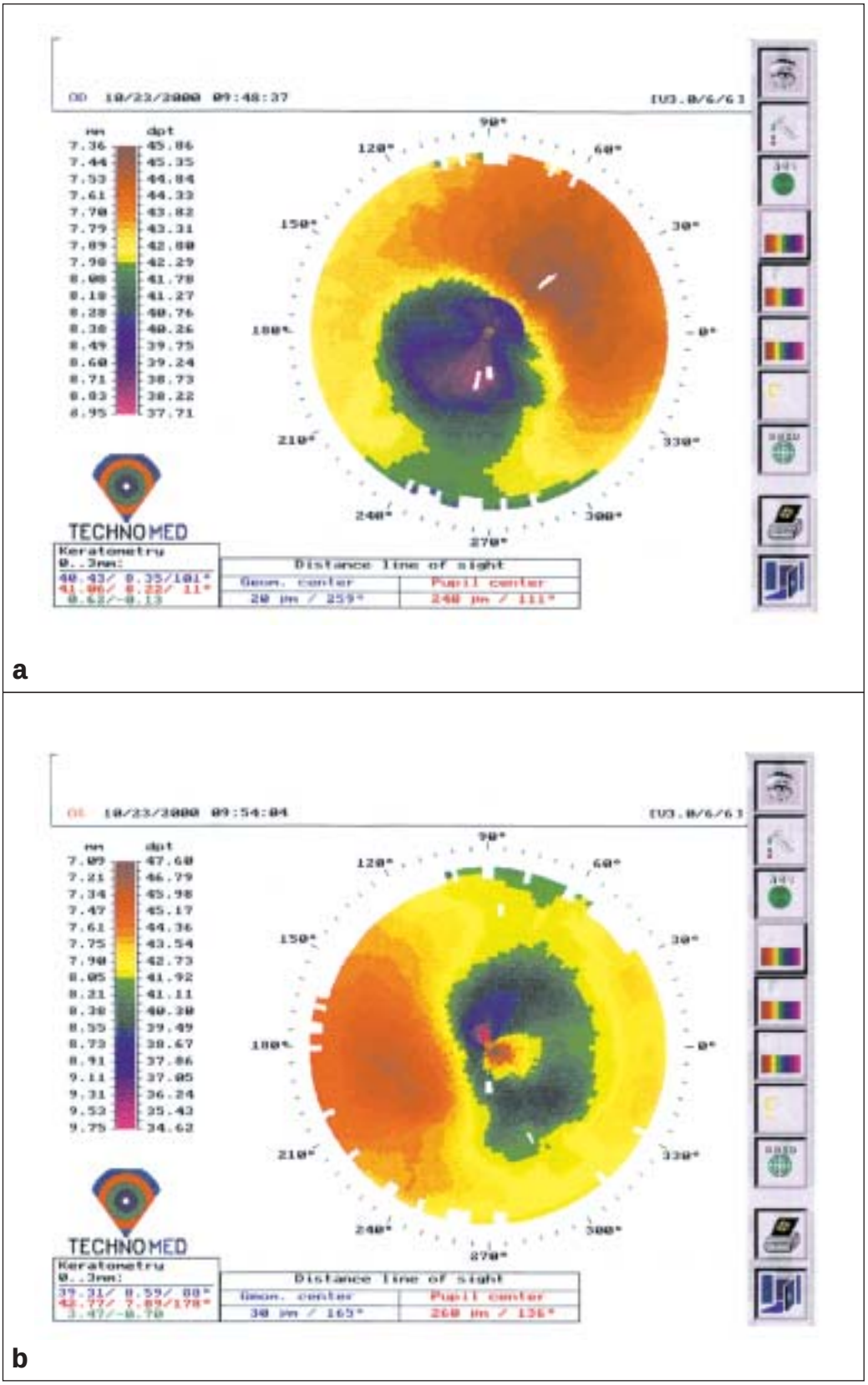

Fig. 3 - *Videokeratography of the right eye shows slightly inferotemporal focal corneal flattening that corresponds to the area of the opacity (**a**); videokeratography of the left eye shows slightly temporal central corneal flattening (**b**).*

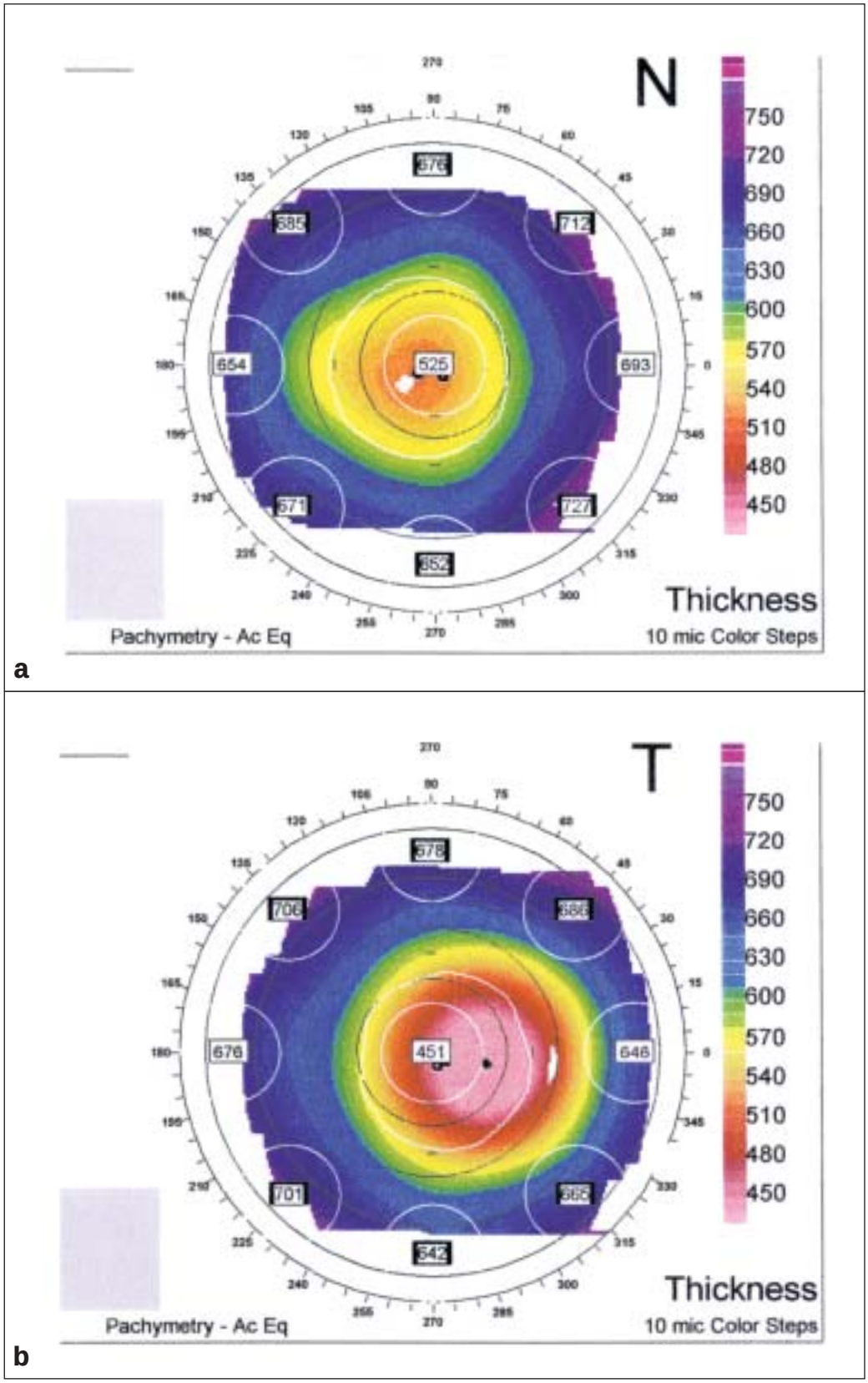

Fig. 4 - *Orbscan pachymetry of the right (**a**) and left (**b**) eyes reveals central thinning in the left eye.*

confined to the interpalpebral fissure, and called it climatic proteoglycan stromal keratopathy. Histopathologic examination of six eyes showed intracellular and extracellular deposits of excess proteoglycans throughout the central stroma. This degeneration occurred in older individuals (mean age 64 years), mainly men. Other corneal degenerations associated with climatic exposure, such as climatic droplet keratopathy and mild calcific band keratopathy, were present in some eyes. Climatic stromal keratopathy has been observed in patients living in Saudi Arabia, and climatic factors (hot, dry climate and solar irradiation and microtrauma from desert sand) were suggested as the probable cause of this corneal degeneration. The mild to moderate corneal haze was reportedly confined to the anterior half to full thickness of the stroma. A similar corneal pathology has not been reported since then.

The clinical appearance of our case closely resembles climatic proteoglycan stromal keratopathy. The frequent finding of focal flattening and thinning of the central cornea overlying the stromal haze, reported by Waring et al, was also present in our case. However, the corneal opacity in our patient was seen earlier and was diagnosed at a younger age and the climatic and environmental factors in our country are not as harsh as in Saudi Arabia. It is not clear whether this bilateral, symmetric corneal disorder in a patient with no affected family members should be described as a dystrophy or degeneration. It is important to report such rarities in order to cast light on new corneal abnormalities.

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